UNMATCHED LEFT PARENTHESIS '(SAPOSIN'
The number of right parentheses in a query must be equal to the number of left parentheses.

=> s saposin (p) blood
L9 44 SAPOSIN (P) BLOOD

=> d ibib abs 1-44

L9 ANSWER 1 OF 44 CAPLUS COPYRIGHT 2004 ACS on STN
ACCESSION NUMBER: 2004:941423 CAPLUS
TITLE: Immunoquantification of α-galactosidase: Evaluation for the diagnosis of fabry disease
AUTHOR(S): Fuller, Maria; Lovejoy, Melanie; Brooks, Doug A.; Harkin, Miriam L.; Hopwood, John J.; Meikle, Peter J.
CORPORATE SOURCE: Lysosomal Diseases Research Unit, Department of Genetic Medicine, Women's and Children's Hospital, North Adelaide, Australia
SOURCE: Clinical Chemistry (Washington, DC, United States) (2004), 50(11), 1979-1985
CODEN: CLCHAU; ISSN: 0009-9147
PUBLISHER: American Association for Clinical Chemistry
DOCUMENT TYPE: Journal
LANGUAGE: English
AB Background: Fabry disease is an X-linked inborn error of glycosphingolipid catabolism resulting from a deficiency of the lysosomal exoglycohydrolase, α-galactosidase. Enzyme replacement therapy is currently available for Fabry disease, but early diagnosis before the onset of irreversible pathol. will be mandatory for successful treatment. Presymptomatic detection would be possible through the use of a newborn-screening program. We report on the use of sensitive assays for the measurement of α-galactosidase protein and activity and for the protein **saposin** C, which are diagnostic markers for Fabry disease. Methods: Two sensitive immunoassays for the measurement of α-galactosidase activity and protein were used to determine the concns. of α-galactosidase in dried filter-paper **blood** spots and plasma samples from control patients and patients with a lysosomal storage disorder (LSD). Results: Fabry hemizygous individuals were clearly identified from control populations by decreases in both α-galactosidase activity and protein. Fabry heterozygotes generally fell between the hemizygotes and controls. Including the measurement of **saposin** C enabled differentiation between Fabry heterozygotes and controls. In **blood** spots, all Fabry individuals could be distinguished from control **blood** spots as well as from 16 other LSD patients. Conclusions: The determination of α-galactosidase activity or protein in dried filter-paper **blood** spots could be used for the diagnosis of Fabry patients. With further validation, these assays could be used for the identification of Fabry patients in newborn-screening programs and may also be suitable for screening high-risk populations.
REFERENCE COUNT: 19 THERE ARE 19 CITED REFERENCES AVAILABLE FOR THIS RECORD. ALL CITATIONS AVAILABLE IN THE RE FORMAT

L9 ANSWER 2 OF 44 CAPLUS COPYRIGHT 2004 ACS on STN
ACCESSION NUMBER: 2004:817730 CAPLUS

Connecting via Winsock to STN

Welcome to STN International! Enter x:x

LOGINID:ssspta1641ayl

PASSWORD:
* * * * * * RECONNECTED TO STN INTERNATIONAL * * * * * *
SESSION RESUMED IN FILE 'CAPLUS, BIOSIS, MEDLINE, EMBASE'
AT 18:10:15 ON 09 DEC 2004
FILE 'CAPLUS' ENTERED AT 18:10:15 ON 09 DEC 2004
COPYRIGHT (C) 2004 AMERICAN CHEMICAL SOCIETY (ACS)
FILE 'BIOSIS' ENTERED AT 18:10:15 ON 09 DEC 2004
Copyright (c) 2004 The Thomson Corporation.
FILE 'MEDLINE' ENTERED AT 18:10:15 ON 09 DEC 2004
FILE 'EMBASE' ENTERED AT 18:10:15 ON 09 DEC 2004
COPYRIGHT (C) 2004 Elsevier Inc. All rights reserved.
COST IN U.S. DOLLARS

	SINCE FILE ENTRY	TOTAL SESSION
FULL ESTIMATED COST	324.93	325.14

DISCOUNT AMOUNTS (FOR QUALIFYING ACCOUNTS)

	SINCE FILE ENTRY	TOTAL SESSION
CA SUBSCRIBER PRICE	-37.80	-37.80

=> file .bio
COST IN U.S. DOLLARS

	SINCE FILE ENTRY	TOTAL SESSION
FULL ESTIMATED COST	324.93	325.14

DISCOUNT AMOUNTS (FOR QUALIFYING ACCOUNTS)

	SINCE FILE ENTRY	TOTAL SESSION
CA SUBSCRIBER PRICE	-37.80	-37.80

FILE 'CAPLUS' ENTERED AT 18:10:27 ON 09 DEC 2004
USE IS SUBJECT TO THE TERMS OF YOUR STN CUSTOMER AGREEMENT.
PLEASE SEE "HELP USAGETERMS" FOR DETAILS.
COPYRIGHT (C) 2004 AMERICAN CHEMICAL SOCIETY (ACS)

FILE 'BIOSIS' ENTERED AT 18:10:27 ON 09 DEC 2004
Copyright (c) 2004 The Thomson Corporation.

FILE 'MEDLINE' ENTERED AT 18:10:27 ON 09 DEC 2004

FILE 'EMBASE' ENTERED AT 18:10:27 ON 09 DEC 2004
COPYRIGHT (C) 2004 Elsevier Inc. All rights reserved.

=> s saposin
L8 1288 SAPOSIN

=> s (saposin (p) (blood or serum or urine or amniotic)
UNMATCHED LEFT PARENTHESIS '(SAPOSIN'
The number of right parentheses in a query must be equal to the
number of left parentheses.

=> s (saposin (p) (blood)

3 FILES SEARCHED...
L10 646 SAPOSIN (1W) (A OR C OR D)

=> s ((saposin) (1w) (a or c or d)) (p) blood
3 FILES SEARCHED...
L11 28 ((SAPOSIN) (1W) (A OR C OR D)) (P) BLOOD

=> d ibib abs 1-28

L11 ANSWER 1 OF 28 CAPLUS COPYRIGHT 2004 ACS on STN
ACCESSION NUMBER: 2004:941423 CAPLUS
TITLE: Immunoquantification of α-galactosidase: Evaluation for the diagnosis of fabry disease
AUTHOR(S): Fuller, Maria; Lovejoy, Melanie; Brooks, Doug A.; Harkin, Miriam L.; Hopwood, John J.; Meikle, Peter J.
CORPORATE SOURCE: Lysosomal Diseases Research Unit, Department of Genetic Medicine, Women's and Children's Hospital, North Adelaide, Australia
SOURCE: Clinical Chemistry (Washington, DC, United States) (2004), 50(11), 1979-1985
CODEN: CLCHAU; ISSN: 0009-9147
PUBLISHER: American Association for Clinical Chemistry
DOCUMENT TYPE: Journal
LANGUAGE: English
AB Background: Fabry disease is an X-linked inborn error of glycosphingolipid catabolism resulting from a deficiency of the lysosomal exoglycohydrolase, α-galactosidase. Enzyme replacement therapy is currently available for Fabry disease, but early diagnosis before the onset of irreversible pathol. will be mandatory for successful treatment. Presymptomatic detection would be possible through the use of a newborn-screening program. We report on the use of sensitive assays for the measurement of α-galactosidase protein and activity and for the protein **saposin C**, which are diagnostic markers for Fabry disease. Methods: Two sensitive immunoassays for the measurement of α-galactosidase activity and protein were used to determine the concns. of α-galactosidase in dried filter-paper **blood** spots and plasma samples from control patients and patients with a lysosomal storage disorder (LSD). Results: Fabry hemizygous individuals were clearly identified from control populations by decreases in both α-galactosidase activity and protein. Fabry heterozygotes generally fell between the hemizygotes and controls. Including the measurement of **saposin C** enabled differentiation between Fabry heterozygotes and controls. In **blood** spots, all Fabry individuals could be distinguished from control **blood** spots as well as from 16 other LSD patients. Conclusions: The determination of α-galactosidase activity or protein in dried filter-paper **blood** spots could be used for the diagnosis of Fabry patients. With further validation, these assays could be used for the identification of Fabry patients in newborn-screening programs and may also be suitable for screening high-risk populations.
REFERENCE COUNT: 19 THERE ARE 19 CITED REFERENCES AVAILABLE FOR THIS RECORD. ALL CITATIONS AVAILABLE IN THE RE FORMAT

L11 ANSWER 2 OF 28 CAPLUS COPYRIGHT 2004 ACS on STN

that this precursor cell in the digesting macrophage system also has an impaired metabolic catabolism for lipopigments (3). Immunohistochemical studies indicate that microglial reaction in NCL brain is limited to resident microglia without contribution by circulating monocytes (4). The granular osmiophilic deposit (GROD) type of NCL has now been established not only in infantile, but also in late-infantile, juvenile, and protracted-juvenile NCL (5). A European Tissue Registry established within the framework of a European Concerted Action on Neuronal Ceroid-Lipofuscinosis may form the basis for additional collaborative studies on NCL, including both biopsy and autopsy tissues.

L9 ANSWER 44 OF 44 EMBASE COPYRIGHT 2004 ELSEVIER INC. ALL RIGHTS RESERVED.
on STN

ACCESSION NUMBER: 90030178 EMBASE
DOCUMENT NUMBER: 1990030178
TITLE: Sphingolipid hydrolase activator proteins and their precursors.
AUTHOR: Sano A.; Hineno T.; Mizuno T.; Kondoh K.; Ueno S.; Kakimoto Y.; Inui K.
CORPORATE SOURCE: Department of Neuropsychiatry, Ehime University School of Medicine, Ehime 791-02, Japan
SOURCE: Biochemical and Biophysical Research Communications, (1989) 165/3 (1191-1197).
ISSN: 0006-291X CODEN: BBRCA
COUNTRY: United States
DOCUMENT TYPE: Journal; Article
FILE SEGMENT: 025 Hematology
029 Clinical Biochemistry
LANGUAGE: English
SUMMARY LANGUAGE: English

AB Activator proteins for sphingolipid hydrolases (**saposins**) are small acidic, heat-stable glycoproteins that stimulate the hydrolysis of sphingolipids by lysosomal enzymes. The molecular mass of each stimulator is about 10 kDa, but glycosylated forms of higher mass exist too. The distribution and developmental changes in two **saposins** and their precursor proteins were studied with the aid of monospecific antibodies against **saposin**-B and **saposin**-C. They show a wide distribution in rat organs and forms intermediate between **saposin** and prosaposin (the precursor protein containing four different **saposin** units) could be seen. The amount of **saposin** and the degree of processing from prosaposin are quite different in different tissues. The **saposins** are the dominant forms in spleen, lung, liver, and kidney, while skeletal muscle, heart, and brain contain mainly precursor forms. In human **blood**, leukocytes contain mainly **saposin**, while plasma contains mainly precursor forms and platelets show many forms. Their subcellular distribution was studied using rat liver. The **saposins** of approximately 20 kDa are dominant in the light mitochondrial, mitochondrial, and microsomal fractions, following the distribution of the activity of a lysosomal marker enzyme. The nuclear fraction exhibits bands corresponding to non-glycosylated **saposin**. The soluble fraction contained much precursor forms. A developmental study of rat brain showed that the concentration of **saposin** precursors increased with age.

=> saposin (1w) (a or c or d)

developed by creating a null allele in embryonic stem cells through gene targeting to investigate the phenotypic diversity of prosaposin mutations and the involvement of this protein in lysosomal storage diseases, and for the development of therapeutic approaches. Mice homozygous mutants die at the age of 35-40 days and neurological disorders contribute to the early demise of the mutant mice. The male reproductive organs in homozygous mutants show several abnormalities, such as a decrease in testis size with reduced spermiogenesis and an involution of the prostate, seminal vesicles, and epididymis. In these animals, the **blood** levels of testosterone remain normal. In the prostate of homozygous mutants, only the basal epithelial cells appear to be present, while the secretory cells are absent. These findings suggest that prosaposin may be involved in the development and maintenance of the male reproductive organs, as well as, in cellular differentiation.

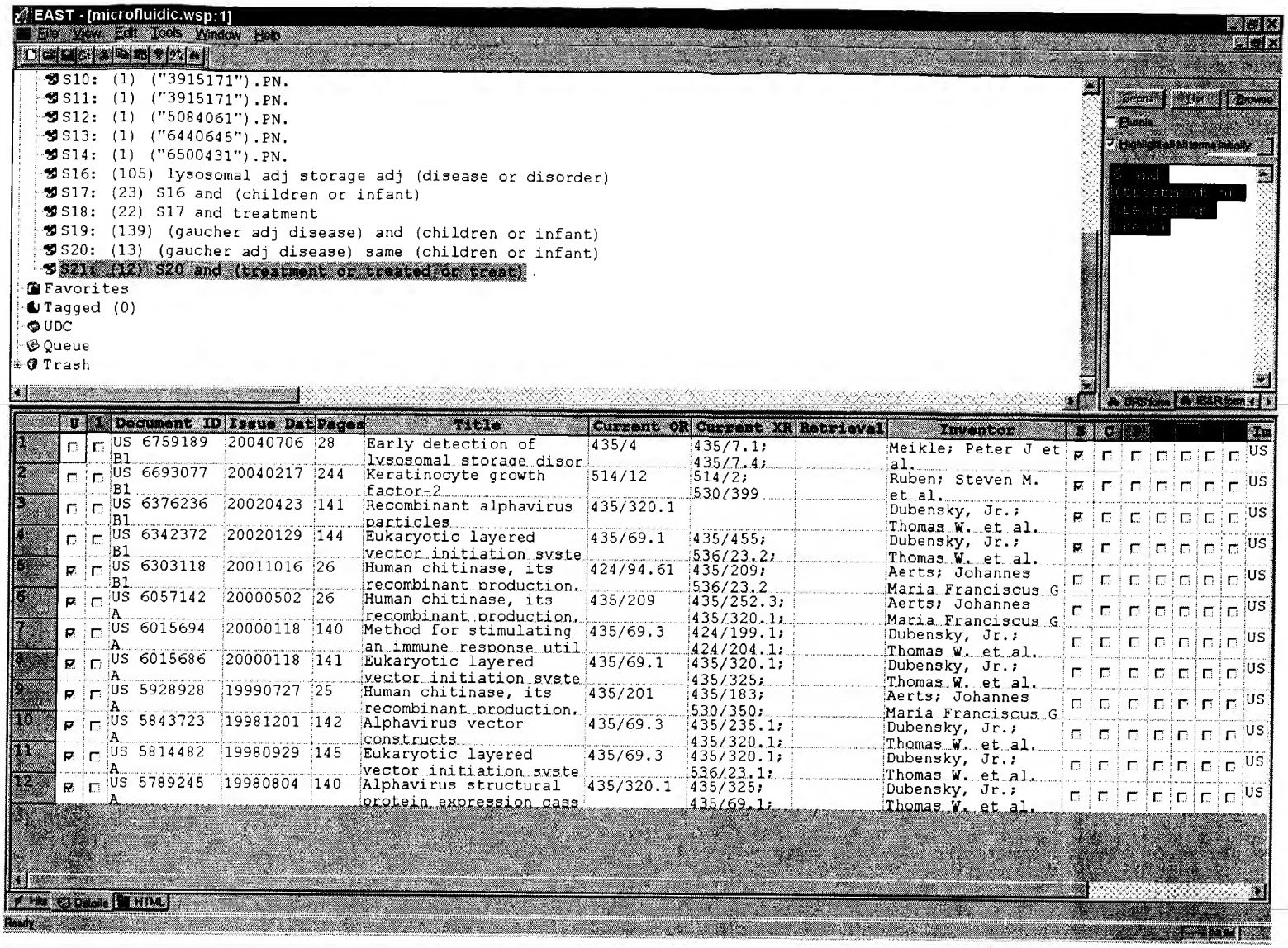
L11 ANSWER 22 OF 28 MEDLINE on STN
ACCESSION NUMBER: 90121224 MEDLINE
DOCUMENT NUMBER: PubMed ID: 2610686
TITLE: Sphingolipid hydrolase activator proteins and their precursors.
AUTHOR: Sano A; Hineno T; Mizuno T; Kondoh K; Ueno S; Kakimoto Y; Inui K
CORPORATE SOURCE: Department of Neuropsychiatry, Ehime University School of Medicine, Japan.
SOURCE: Biochemical and biophysical research communications, (1989 Dec 29) 165 (3) 1191-7.
Journal code: 0372516. ISSN: 0006-291X.
PUB. COUNTRY: United States
DOCUMENT TYPE: Journal; Article; (JOURNAL ARTICLE)
LANGUAGE: English
FILE SEGMENT: Priority Journals
ENTRY MONTH: 199002
ENTRY DATE: Entered STN: 19900328
Last Updated on STN: 19900328
Entered Medline: 19900213

AB Activator proteins for sphingolipid hydrolases (saposins) are small acidic, heat-stable glycoproteins that stimulate the hydrolysis of sphingolipids by lysosomal enzymes. The molecular mass of each stimulator is about 10 kDa, but glycosylated forms of higher mass exist too. The distribution and developmental changes in two saposins and their precursor proteins were studied with the aid of monospecific antibodies against saposin-B and **saposin-C**. They show a wide distribution in rat organs and forms intermediate between saposin and prosaposin (the precursor protein containing four different saposin units) could be seen. The amount of saposin and the degree of processing from prosaposin are quite different in different tissues. The saposins are the dominant forms in spleen, lung, liver, and kidney, while skeletal muscle, heart, and brain contain mainly precursor forms. In human **blood**, leukocytes contain mainly saposin, while plasma contains mainly precursor forms and platelets show many forms. Their subcellular distribution was studied using rat liver. The saposins of approximately

20 kDa are dominant in the light mitochondrial, mitochondrial, and microsomal fractions, following the distribution of the activity of a lysosomal marker enzyme. The nuclear fraction exhibits bands corresponding to non-glycosylated saposin. The soluble fraction contained much precursor forms. A developmental study of rat brain showed that the concentration of saposin precursors increased with age.

L11 ANSWER 23 OF 28 EMBASE
on STN

ACCESSION NUMBER: 2004460372 EMBASE
TITLE: Immunoquantification of α-galactosidase: Evaluation for the diagnosis of fabry disease.
AUTHOR: Fuller M.; Lovejoy M.; Brooks D.A.; Harkin M.L.; Hopwood J.J.; Meikle P.J.
CORPORATE SOURCE: M. Fuller, Lysosomal Diseases Research Unit, Department of Genetic Medicine, Women's and Children's Hospital, 72 King William Rd., North Adelaide, SA 5006, Australia. maria.fuller@adelaide.edu.au
SOURCE: Clinical Chemistry, (2004) 50/11 (1979-1985).
Refs: 19
ISSN: 0009-9147 CODEN: CLCHAU
COUNTRY: United States
DOCUMENT TYPE: Journal; Article
FILE SEGMENT: 007 Pediatrics and Pediatric Surgery
029 Clinical Biochemistry
LANGUAGE: English
SUMMARY LANGUAGE: English
AB Background: Fabry disease is an X-linked inborn error of glycosphingolipid catabolism resulting from a deficiency of the lysosomal exoglycohydrolase, α-galactosidase. Enzyme replacement therapy is currently available for Fabry disease, but early diagnosis before the onset of irreversible pathology will be mandatory for successful treatment. Presymptomatic detection would be possible through the use of a newborn-screening program. We report on the use of sensitive assays for the measurement of α-galactosidase protein and activity and for the protein **saposin C**, which are diagnostic markers for Fabry disease. Methods: Two sensitive immunoassays for the measurement of α-galactosidase activity and protein were used to determine the concentrations of α-galactosidase in dried filter-paper **blood** spots and plasma samples from control patients and patients with a lysosomal storage disorder (LSD). Results: Fabry hemizygous individuals were clearly identified from control populations by decreases in both α-galactosidase activity and protein. Fabry heterozygotes generally fell between the hemizygotes and controls. Including the measurement of **saposin C** enabled differentiation between Fabry heterozygotes and controls. In **blood** spots, all Fabry individuals could be distinguished from control **blood** spots as well as from 16 other LSD patients. Conclusions: The determination of α-galactosidase activity or protein in dried filter-paper **blood** spots could be used for the diagnosis of Fabry patients. With further validation, these assays could be used for the identification of Fabry patients in newborn-screening programs and may also be suitable for screening high-risk populations. .COPYRGT. 2004 American Association for Clinical Chemistry.



	U	1	Document ID	Issue Dat	Pages	Title	Current OR	Current XR	Retrieval	Inventor	S	C							In
1	☐	☐	US 6759189 B1	20040706	28	Early detection of lysosomal storage disor	435/4	435/7.1; 435/7.4;		Meikle; Peter J et al.	☑	☐	☐	☐	☐	☐	☐	☐	US
2	☐	☐	US 6693077 B1	20040217	244	Keratinocyte growth factor-2	514/12	514/2; 530/399		Ruben; Steven M. et al.	☑	☐	☐	☐	☐	☐	☐	☐	US
3	☐	☐	US 6376236 B1	20020423	141	Recombinant alphavirus particles	435/320.1			Dubensky, Jr.; Thomas W. et al.	☑	☐	☐	☐	☐	☐	☐	☐	US
4	☐	☐	US 6342372 B1	20020129	144	Eukaryotic layered vector initiation syste	435/69.1	435/455; 536/23.2;		Dubensky, Jr.; Thomas W. et al.	☑	☐	☐	☐	☐	☐	☐	☐	US
5	☑	☐	US 6303118 B1	20011016	26	Human chitinase, its recombinant production,	424/94.61	435/209; 536/23.2		Aerts; Johannes Maria Franciscus G	☐	☐	☐	☐	☐	☐	☐	☐	US
6	☑	☐	US 6057142 A	20000502	26	Human chitinase, its recombinant production,	435/209	435/252.3; 435/320.1;		Aerts; Johannes Maria Franciscus G	☐	☐	☐	☐	☐	☐	☐	☐	US
7	☑	☐	US 6015694 A	20000118	140	Method for stimulating an immune response util	435/69.3	424/199.1; 424/204.1;		Dubensky, Jr.; Thomas W. et al.	☐	☐	☐	☐	☐	☐	☐	☐	US
8	☑	☐	US 6015686 A	20000118	141	Eukaryotic layered vector initiation syste	435/69.1	435/320.1; 435/325;		Dubensky, Jr.; Thomas W. et al.	☐	☐	☐	☐	☐	☐	☐	☐	US
9	☑	☐	US 5928928 A	19990727	25	Human chitinase, its recombinant production,	435/201	435/183; 530/350;		Aerts; Johannes Maria Franciscus G	☐	☐	☐	☐	☐	☐	☐	☐	US
10	☑	☐	US 5843723 A	19981201	142	Alphavirus vector constructs	435/69.3	435/235.1; 435/320.1;		Dubensky, Jr.; Thomas W. et al.	☐	☐	☐	☐	☐	☐	☐	☐	US
11	☑	☐	US 5814482 A	19980929	145	Eukaryotic layered vector initiation syste	435/69.3	435/320.1; 536/23.1;		Dubensky, Jr.; Thomas W. et al.	☐	☐	☐	☐	☐	☐	☐	☐	US
12	☑	☐	US 5789245 A	19980804	140	Alphavirus structural protein expression cass	435/320.1	435/325; 435/69.1;		Dubensky, Jr.; Thomas W. et al.	☐	☐	☐	☐	☐	☐	☐	☐	US